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Small Bowel Leiomyosarcoma Presenting with Jejuno-jejunal Intussusception: A Case Report and Literature Review

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Abstract: Primary gastrointestinal (GI) sarcomas in general are a very rare entity. Leiomyosarcoma (LMS) is the most common histiotype of this group of malignant neoplasms. Intestinal LMS are mesenchymal tumors of smooth muscle origin which occur mainly in the fifth and sixth decades of life. Abdominal pain and GI bleeding are the most common common presentation, but some of them present with intestinal obstruction. We present a case of an 80-year-old male who presented to us with acute intestinal obstruction, with otherwise unremarkable past medical history. CT scan thorax/abdomen/pelvis showed distal jejunal intussusception with a small bowel lesion. He underwent laparotomy which revealed a small bowel intussusception, with a 5cmx4cmx2cm pedunculated intra-luminal tumour as the leading point. Small bowel resection and end to end anastomosis were done. HPE confirmed a poorly differentiated leiomyosarcoma, with expression of Vimentin, Caldesmon and SMA in the immunohistochemistry. Early diagnosis of small bowel LMS remains a challenge considering the non-specific or sub-acute symptoms. Although CT scan is useful in confirming an anatomical abnormality, final diagnosis requires histopathological analysis. The treatment of such tumours remains predominantly centered on surgical resection and prognosis is dependent on tumour size and histological staging.

Keywords: Leiomyosarcoma, intestinal obstruction, intussusception, immunohistochemistry

INTRODUCTION

Small bowel tumours include a heterogenous group of benign and malignant lesions. The most common malignant lesions are adenocarcinoma, neuroendocrine tumours, sarcomas and lymphomas but more than forty different histological subtypes are described [1]. Primary gastrointestinal (GI) sarcomas in general are a very rare entity, accounting for 1-2% of GI malignancies. Of this uncommon group of highly malignant neoplasms, the leiomyosarcoma (LMS) is the most common histiotype[2].

Intestinal LMS are mesenchymal tumors of smooth muscle origin. They occur mainly in the fifth and sixth decades of life, and abdominal pain and GI bleeding have been reported to be the most common clinical signs at presentation [3]. Spectrum of symptoms of small bowel tumours differs slightly from the other parts of GI tract. Other than abdominal pain and GI bleeding, weight loss, intestinal obstruction (including intussusception), presence of abdominal mass, jaundice and perforation are also known symptoms associated to malignant small bowel tumours[4].

We presented to you a rare case of small bowel leiomyosarcoma presenting with jejuno-jejunal intussusception as a case report.

CASE PRESENTATION

An 80-year-old gentleman who was a chronic smoker came to emergency department presented with generalized colicky abdominal pain for three days duration, associated with no bowel opening and emesis. On further questioning, he had a two months history of chronic cough, and was treated once for pneumonia one month before the abdominal complaints. He otherwise had no alteration of bowel habit. The medical history and family history were both unremarkable.

On initial examination, he appeared cachexic, not jaundiced and generally well. Abdominal examination revealed a tender mass over the left side of the lower abdomen. Per rectal examination, lungs and cardiovascular examination were unremarkable.

Laboratory investigation showed microcytic hypochromic anemia with a hemoglobin level of 74.0 g/L. No abnormalities were observed in leucocytes,

thrombocyte, liver enzyme, renal profile, electrolytes and coagulation profile.

Abdominal x-ray revealed dilated small bowel. Subsequently a computed tomography (CT) scan of Abdomen, Pelvis and Thorax was done and revealed a distal jejunal intussusception causing small bowel dilatation, a small bowel lesion at the left lumbar region with left adrenal mass. There were also lung masses with supraclavicular and mediastinal lymphadenopathy highly suggestive of metastatic disease.

An urgent laparotomy was performed which revealed a small bowel intussusception of 20 cm length at 16 cm from duodeno-jejunal junction with reactive lymph nodes palpable over the mesentery of the intussuscepted part. The small bowel proximal to the intussusception was dilated and the bowel distal to it was collapsed. There was mild ascites, and multiple nodules were palpable over the lateral side of the left lobe of liver. The rest of small bowel, stomach, colon and rectum were normal with no omental nodes. The intussusception was bivalve-opened and revealed a 5cm x 4cm x 2cm pedunculated tumour as the leading point of the intussusception. The intussusception was resected and primary bowel anastomosis was done. Unfortunately, post-operatively, he developed septicemia and myocardial infarction and succumbed five days following surgery.

Gross pathology analysis showed pedunculated tumour measures 5cm x 4 cm x 2 cm, with surface erosion and haemorrhage seen. Cut surface shows homogenous whitish mass, and the tumour is noted to arise from submucosal layer.

The microscopic appearance shows features of poorly differentiated leiomyosarcoma. Immunohistochemistry noted; the tumour cells express Vimentin, Clemson and SMA. Pan CK show focal positivity. They do not express EMA, CK7, TTF-1, Desman, CD117, DOG1, S100 and CD34. Proliferative index (Ki67) is high.

DISCUSSION

Tumours of small bowel account for less than 5% of all GI malignancies. Its incidence is 22.7 cases per million with sarcomas accounting for 1.2% of these [5]. Leiomyosarcomas of the small bowel are therefore extremely rare entities to the extent that World Health Organization can provide no meaningful data on demographic or clinical features as result. Only 26 cases of small bowel leiomyosarcomas were identified following the advent of its robust differentiation from gastrointestinal stromal tumors (GISTs), illustrating the importance of reporting such cases [6].

Small bowel tumours are usually asymptomatic at early stages, and difficult to visualize by upper and lower endoscopy. Its diagnosis can prove difficult; as the presentation can be delayed until metastases are present [1]. Among all small bowel tumours, 83% of the tumors caused symptoms and were diagnosed at operation [4]. They commonly present with abdominal pain, GI bleeding, weight loss, intestinal obstruction (including intussusception), presence of abdominal mass, jaundice or perforation. Obstruction occurred in 16% of cases and was manifested as intussusception in only 3.2% of cases [4].

Intussusception is an invagination of a segment of the gastrointestinal tract into an adjacent segment. Intussusception is rare in adults accounting for one percent of all cases of intestinal obstruction and five percent of all intussusceptions [8]. In adults, intussusception is typically due to a lead point within the bowel, which is malignant in over 50% of cases [9].

Diagnosis of small bowel tumour is difficult and the optimum technique varies depending on the site and size of the tumour. Upper GI radiographic methods including small bowel follow through (SBFT), computed tomography (CT), enterocolitis & heterography may reveal intestinal or lymph node masses, mucosal defects and sometimes intussusception [10]. CT scanning has an important role in evaluating a primary tumour, determining local invasion of surrounding structures and identifying lymph nodes and distal metastasis although peritoneal metastases are sometimes difficult to observe radiologically [11].

Despite advances in imaging, the differentiation between benign and malignant tumours remains extremely difficult pre-operatively [12]. The aetiology of many cases does not become apparent until after definitive resection [12], the case presented here being a prime example.

Histologically, the tumour demonstrates spindle cells with cigar shaped nuclei with prominent cytologic atypia and mitotic figures. Leiomyosarcomas are differentiated from GISTs by the lack of CD117(c-KIT), DOG1 and CD34 as well as presence of smooth muscle actin (SMA) and desmin[6,13]. The tumour can be graded using Trojani[15] and French[16] systems for soft tissue sarcomas, denoting its aggression.

The treatment of all small bowel tumours remains centered around radical surgical resection [7]. It is important to differentiate GISTs from leiomyosarcomas as the management of these tumours are very different. Localized GIST and non-GIST sarcomas are managed similarly to other tumours with margin negative surgical resection being the primary goal. Sarcomas rarely metastasize to lymph nodes so extensive lymphadenectomy is not necessary [1]. Whilst adjuvant chemotherapy agents are in use for GISTs [13] and uterine [14] leiomyosarcomas, their use has not been proven in the case of small bowel disease [12].

With curative resection, the 5-year survival rate for patients with leiomyosarcoma of the small intestine is 40-50%. Distant metastasis decreases the survival rate to around 30% [13]. However, given the proportion of cases with late presentations, metastases are an unfortunately common finding, hence the overall poor prognosis [12].

CONCLUSION

Small bowel LMS with a focus of jejuno-jejunal intussusception as presentation is a very rare entity, thus the importance of reporting such case. It may present with a variety of symptoms, including intestinal obstruction. Early diagnosis remains a challenge considering the non-specific or sub-acute symptoms. Although CT scan is useful in confirming an anatomical abnormality, final diagnosis requires histopathological analysis. Histopathologically, they are differentiated from GISTs by the lack of CD117(c-KIT), DOG1 and CD34 as well as presence of smooth muscle actin (SMA) and desmin. The treatment of such tumours remains predominantly centered around surgical resection, and prognosis is dependent on tumour size and histological staging.

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